# CIRCULATORY FACTORS AFFECTING ANESTHESIA IN SURGERY FOR CONGENITAL HEART DISEASE *

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THE special problems involved in the anesthetic management of patients with congenital heart disease usually are the direct result of altered hemodynamics and characteristic types of inefficiency imposed upon the heart by virtue either of abnormal shunts or of anomalous obstructions in the heart or great vessels (1, 2). The best preparation for the anesthetist in the management of these difficulties is a thorough understanding of the abnormal physiology of the patient coming to operation and the expected effect of the operation upon this unnatural physiology. Consequently, the chief purpose of this paper will be to review the basic physiology in the three types of congenital cardiac defects most commonly treated surgically, namely; patent ductus arteriosus, tetralogy of Fallot and coarctation of the aorta. With a thorough understanding of the underlying anatomy, the resultant hemodynamic and biochemical changes and the surgical procedures involved, the anesthetist will find logical answers to most of his problems, and will be able to prevent the occurrence of foreseen difficulties.

## PATENT DUCTUS ARTERIOSUS

In order to obtain a clear picture of the patent ductus arteriosus, one does well to review its function in the fetal circulation (fig. 1). Here one sees the oxygenated blood coming to the fetus by way of the umbilical vein, mixing with reduced blood in the portal vein and inferior vena cava and entering the right auricle. Most of this blood from the inferior vena cava passes along the posterior aspect of the right auricle, thence through the foramen ovale into the left atrium, left ventricle and out through the aorta, so that the head and upper limbs receive the better oxygenated blood (3). The venous blood returning by way of the superior vena cava to the right auricle passes to the right ventricle and then up through the pulmonary artery. Here, since the lungs are not functioning, little blood passes out through the pulmonary vessels to those structures. The pressure in the pulmonary

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circuit at this time exceeds that in the aorta; consequently it will be noted that the shunted blood flows through the ductus arteriosus in a direction *away* from the heart, from the pulmonary artery into the aorta. From here it proceeds to supply the lower part of the body and lower limbs with oxygenated blood.

In normal infants, closure of the foramen ovale and obliteration of the ductus arteriosus is completed during the first or second month of life (4). Subsequently in these normal individuals, the systemic side of the circulatory system builds up considerable pressure, amounting to about 100 mm. systolic and 60 mm. diastolic, while in the pulmonary artery the pressure remains low and stable, on the average about 28 mm. systolic and 7 mm. diastolic (5).

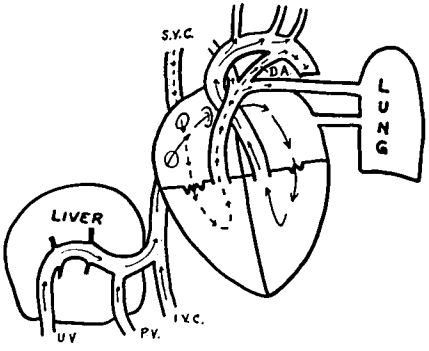



FIG. 1. Fetal circulation: u.v., umbilical vein; p.v., portal vein; i.v.c., inferior vena cava; s.v.c., superior vena cava; d.a., ductus arteriosus. Ductus arteriosus shunts blood from pulmonary artery into aorta, away from the heart.

Should the ductus arteriosus fail to close, the resulting physiologic abnormalities characteristic of the "patent ductus arteriosus" become evident (fig. 2). These abnormalities are most entirely hemodynamic changes, as will be seen. At birth, the blood pressure in the aorta becomes slightly greater than that in the pulmonary artery. This causes a reversal in the direction of the flow of blood through the ductus. From then on, the blood flows from the aorta into the pulmonary artery in a direction *toward* the heart.

At first the difference in pressure is not great enough to cause a significant amount of blood to flow through the shunt, and consequently no murmur is heard in these patients in early infancy. Later, however, with the development of more peripheral resistance in the systemic circulation, an ever-increasing amount of blood is shunted through the ductus, and a murmur is heard, first during systole only and later

during both systolic and diastolic phases of the cycle. The resultant continuous murmur gives evidence that even in diastole the aortic pressure is considerably higher than the pulmonic pressure.

It should be mentioned that although there is an interchange of arterial with venous blood, cyanosis is virtually *never* seen in patients with patent ductus arteriosus. This is true because, except under terminal conditions, aortic pressure remains sufficiently higher than pulmonic to prevent any venous blood from entering the aorta (6).

The amount of blood that passes through the patent ductus from aorta to pulmonary artery would not be expected to be great, if one

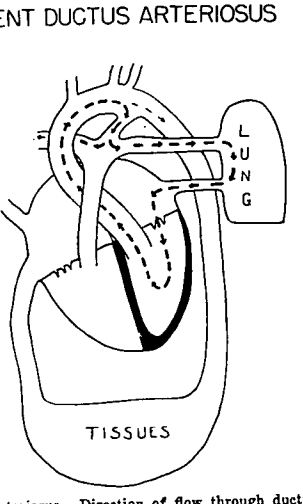



FIG. 2. Patent ductus arteriosus. Direction of flow through ductus is reversed, and recirculation of oxygenated blood through lesser circulation is set up, adding to work of left heart. Cyanosis is not seen in this condition.

were to judge only by the relatively small diameter of the ductus when compared to that of the aorta. It must be borne in mind, however, that blood flowing down the aorta faces a high resistance, while that in the pulmonary system meets but little. Consequently, it will be found that a surprisingly large proportion of the left ventricular output is shunted through the ductus back into the pulmonary circulation. According to the studies of Eppinger, Burwell and Gross (6), as much as 75 per cent of the left ventricular output may pass through the ductus to be wastefully returned to the pulmonary circulation. Here this already oxygenated blood again passes through the lungs

without having performed any oxidative function. This *recirculation of oxygenated blood through the lesser circulation without reaching systemic capillaries is characteristic of the patent ductus arteriosus.*

The existence of an appreciable arteriovenous shunt results in other important changes in blood pressure relationships. The shunt acts as a large leak in an otherwise intact systemic circulation, thereby causing a marked decrease in peripheral resistance. During systole the heart is able to build up arterial pressure to normal, but in diastole the pressure suddenly falls, far below normal. As a result of this lowered diastolic pressure there is the widened pulse pressure typical of this lesion. In general, the systolic pressure will be 100 to 110 mm. systolic, and 50 to 60 mm. diastolic, giving a pulse pressure of 40 or greater in most instances (fig. 3). In the systemic circulation the low diastolic pressure produces a resultant quick, water-hammer type pulse. The effect of the shunt on the pulmonary circulation is to raise the pressure

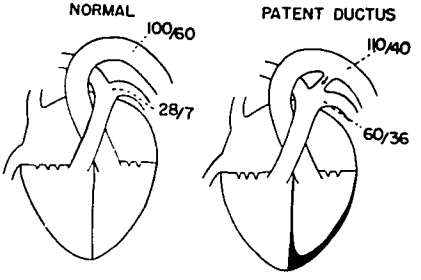



FIG. 3. Effect of patent ductus artesiosus on blood and pulse pressure. Pulmonary arterial pressures are increased. In the systemic circulation the diastolic pressure is decreased, producing widening of pulse pressure.

markedly in the pulmonary arteries. This pulmonary hypertension is evident in the consequent engorgement of these vessels and the hilar vessels of the lungs.

The effect of this shunt upon the heart itself is most important and sets the stage for most of the problems that arise to trouble the anesthetist during these operations.

Since more than half of the left ventricular output may be shunted back into the pulmonary circulation, the left ventricle must increase the total output enough to make up for this inefficiency. To do this the rate increases, as does the output per stroke. A rapid, actively pounding heart develops, and this is followed by left ventricular hypertrophy in the more severe cases. Further demands upon the heart are evidenced in increased irritability of the heart and the appearance of arrhythmias, and finally the development of left ventricular failure.

The clinical picture of the patient as he comes to operation is easily visualized. If the ductus is small, the patient appears essentially normal. With a large shunt, the patient exhibits a proportional degree of underdevelopment. Although the child's color will be good, his heart may be seen to be beating rapidly and forcefully against his thoracic cage, even when the child is at rest. The typical wide pulse pressure will be found.

The purpose of the operation is simply to obliterate the shunt, and thereby to re-establish normal hemodynamics. This may be accomplished either by ligating (7, 8) or by dividing (9, 10) the ductus

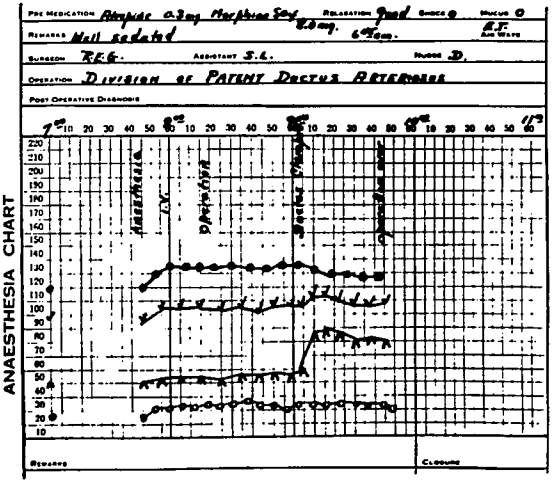



FIG. 4. Anesthesia chart from operation for division of patent ductus arteriosus. Typical reduction of width of pulse pressure is seen at time of clamping of the ductus.

arteriosus. The immediate result of either procedure is the same. The leak in the systemic circulation is stopped, and at once there is a marked rise in diastolic pressure and a resultant reduction in the pulse pressure. This is usually evident in the anesthesia chart (fig. 4). The heart is now able to work efficiently, the rate slows and the beat becomes less forceful. It can be seen that the heart is distended less markedly as the stroke volume diminishes. The pulmonary vessels no longer receive an excessive quota of blood from the aorta, so that the pulmonary pressure returns to normal and engorgement disappears. The unstable, irritable heart soon regains its stability and strength and

shortly afterward the patient begins to make up the developmental deficit that had existed.

In general, surgical correction of patent ductus arteriosus is not frequently complicated by serious difficulties. The operation does not impose a great strain upon the patient, nor does it consume an excessive amount of time. Even when the surgeon divides the ductus, the operation often requires less than two hours. At our institution the mortality in over 455 operations has been slightly more than 1 per cent (November 1, 1950).

As can easily be deduced, problems that do arise center chiefly about patients with unusually large shunts, who have signs of heart strain or failure. These patients should be medicated sufficiently to prevent excitement at the time of induction. Pentothal or nitrous-oxide-oxygen induction followed by ether maintenance has proved satisfactory, although many agents have been used with equally good results (11, 12). We have not employed agents for the specific prevention of cardiac arrhythmias unless a definite arrhythmia existed preoperatively which indicated such medication.

Occasionally patients appear to become worse after the chest is opened. The pulse rate may increase considerably or irregularities of rhythm may appear. The anesthetist may be faced with the difficult decision as to whether or not he should call for abandonment of the operation. Several experiences of this nature have convinced us that whenever possible the surgeon should attempt to carry on as expeditiously as possible, and close off the shunt. By doing this, he will, in all probability, eradicate the actual cause of the trouble, and such palliative agents as pronestyl, procaine or quinidine will not be required. The heart will be able to do its own work efficiently, and need no further assistance. By the time the chest is closed it will be noticed that the pulse has become appreciably slower and less active, and the previously bounding apical impulse is no longer evident.

The postoperative course of these patients is singularly free of complications. Except for an occasional collection of pleural fluid, these children have shown no postoperative problems worthy of mention.

### TETRALOGY OF FALLOT

There are several different congenital cardiac defects which cause cyanosis. That most frequently seen is the syndrome known as the tetralogy of Fallot. The four anatomic defects comprising the tetralogy of Fallot are pulmonary stenosis, overriding of the aorta, interventricular septal defect and right ventricular hypertrophy (2). The first three of these features are primary defects which by imposing an increased load upon the right heart give rise to hypertrophy of the right ventricle, the fourth anatomic defect (fig. 5).

The above anatomic abnormalities bring about two derangements in

the circulation, each of which by itself leads to hypoxia and cyanosis (fig. 6). The narrowing of the pulmonary artery obstructs the usual flow of venous blood from right ventricle to the lungs. Consequently, a decreased amount of reduced blood is returned to the lungs for oxygenation. The severity of the resultant hypoxia varies with the degree of narrowing present in the pulmonary artery.

The second abnormal mechanism is produced by the combined effect of the ventricular septal defect and the overriding aorta, which allows venous blood from the right ventricle to escape into the aorta and mix with the oxidized blood of the systemic circulation. By virtue

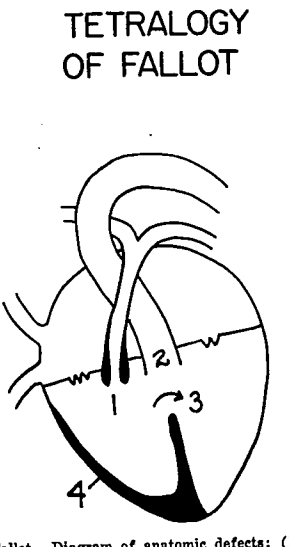



FIG. 5. Tetralogy of Fallot. Diagram of anatomic defects: (1) stenosis of pulmonary artery; (2) aorta overriding ventricular septum; (3) interventricular septal defect and (4) the resultant right ventricular hypertrophy.

of this arteriovenous shunt *a recirculation of blood through the systemic circuit is set up,* whereby the same blood can go to the tissues many times without passing through the lungs (fig. 7). The presence of unoxidized blood in the systemic circulation naturally causes hypoxia and increases the cyanotic tendency already existing. The degree of hypoxia and cyanosis contributed by this mechanism increases with the amount of overriding of the aorta.

Typical hemodynamic alterations occur as a result of these circulatory abnormalities. The obstruction to pulmonary circulation

causes marked hypotension in the lesser circulation. Instead of the normal pulmonary arterial pressure of about 28 mm. systolic and 7 mm. diastolic, the pulmonary arterial pressure in patients with tetralogy of Fallot may be as low as 10 mm. systolic and 7 mm. diastolic. On the systemic side, the blood pressure is usually low also, in the range of 90 mm. systolic and 78 mm. diastolic. The narrow pulse pressure is especially characteristic in these patients. The reason for this has not been clearly defined, but chronic myocardial hypoxia and increased viscosity of the blood are undoubtedly contributing factors.

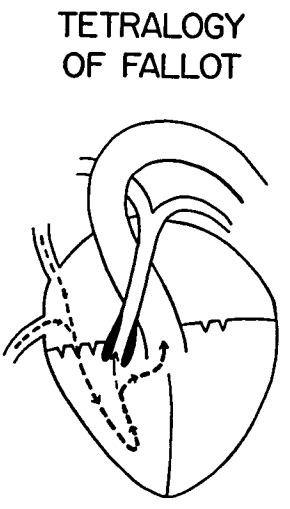



FIG. 6. Abnormal circulation in tetralogy of Fallot: (1) obstruction to blood entering pulmonary vein; (2) venous blood shunted into systemic circulation. Both mechanisms lead to anoxia and cyanosis. Operation partially corrects first, does not alter second defect.

The shunting of blood from the right ventricle into the systemic circulation increases the volume of blood in that circuit at the expense of the blood volume of the lesser circulation, so that there may be twice as much blood going to the tissues as to the lungs. All the blood going to the tissues must return to the right ventricle, consequently the right ventricle will have double the load of the left, and the balance between left and right heart will be upset.

These hemodynamic factors are almost exactly the reverse of those previously described in patients with patent ductus arteriosus who have shunting of blood into the pulmonary artery, resultant pulmonary hypertension, wide pulse pressure and recirculation of

oxidized blood through lungs, rather than recirculation of unoxidized blood to tissues as seen in patients with tetralogy of Fallot.

In patients with tetralogy of Fallot the composition of the blood undergoes important changes in compensation for the defects in the process of oxygenation. In these patients there are often 7,000,000 to 9,000,000 red blood cells per cubic centimeter and hemoglobin content proportionally increases to 18 to 22 gm. per 100 cc. The polycythemia and associated increase in hemoglobin content provide much greater oxygen capacity, thus tending to overcome, in part, the decrease in oxygen saturation. One serious disadvantage is involved here, however, in that this increase in circulating erythrocytes causes a marked

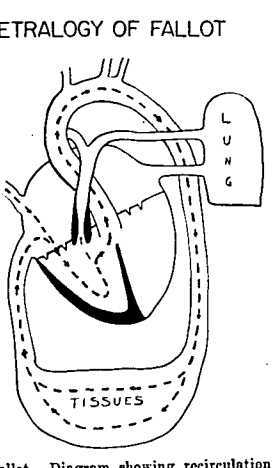



FIG. 7. Tetralogy of Fallot. Diagram showing recirculation of reduced blood through systemic circulation without passage through lungs. Anoxia and cyanosis result. An increased proportion of blood volume remains in systemic circulation, throwing extra load on right ventricle.

hemoconcentration, the hematocrit level often reaching 70 per cent or more. There is a resultant danger of thrombosis of cerebral or other vessels which is ever-present in these patients.

The problems that arise in dealing with these patients, then, revolve around two principal factors: thrombosis and anoxia. The danger of thrombosis is especially marked in the preoperative phase when it is customary to restrict fluids. The resultant dehydration would invite hemoconcentration and thrombosis in the cyanotic patients; consequently, it is important that fluids by oral or intravenous route be continued through the entire preoperative period.

The problem of anoxia is always present, varying only in degree.

When seen on the ward, the patient gives the appearance of being a poor candidate for anesthesia. Deeply cyanosed, with clubbed fingers, blood pressure barely audible, narrow pulse pressure and little or no cardiac reserve, the child gives cause for serious concern, regarding how he will withstand anesthesia and operation if his condition is already precarious. Another complication is the extreme youth of some patients, since in the most severely handicapped, operation may be necessary in infancy. To estimate the patient as an operative risk, several standards may be used, but actually, the *patient's tolerance to excercise* will give the best criterion of his cardiac reserve (13, 14, 15). To the relief of the anesthetist, patients usually improve with preoperative sedation owing to lowering of metabolic activity. Adequate premedication is of great importance in these patients, and morphine, in combination with nembutal and atropine, proves effective for this purpose. McQuiston (16) has reduced oxygen requirement in cyanotic patients by lowering the body temperature. Elevation of temperature certainly should be avoided.

The anesthetic management of patients undergoing surgical intervention for tetralogy of Fallot has been well described by Harmel and Lamont (17), McQuiston (16), and Berger (18, 19). It is essential that excitement and anoxic phenomena, such as spasm and retching, be avoided during induction. To diminish any tendency toward anoxia at this time it is wise to administer oxygen to these children for some moments before inducing anesthesia.

Induction may be accomplished easily by injecting sodium pentothal into a previously started intravenous infusion, while cyclopropane or ethylene may be used in well-sedated children. In our series, ether has been used as the principal agent for maintenance because of its stimulating effect on the myocardium.

The establishment of full surgical anesthesia may further improve the color and general condition of the patient. From the very beginning, however, the anesthetist must be on the lookout for generalized vasomotor depression. It is characteristic of these patients to come to operation with low blood pressure and extremely poor pulse pressure. A blood pressure of 76 mm. systolic and 72 mm. diastolic is not an infrequent finding. For this reason, depressant anesthetic agents or combinations of agents, such as large doses of avertin, curare and morphine, as advocated by Harris (12), would appear to invite increased hazards.

To combat low blood and pulse pressures, measures should be taken early in the procedure. Blood should be administered, and if not effective in maintaining blood pressure, sustaining doses of ephedrine or other vasopressor drugs may be indicated soon after the operation is begun. In our series, the prevention of vasomotor depression has been of great importance in reducing the mortality which at first accompanied this operation.

Once surgical anesthesia has been attained, the anesthetic course will usually be satisfactory until the chest is opened. At this point, collapsing the lung may be extremely hazardous, since it may create just enough additional anoxia to make the situation unbearable for the patient. Consequently, the anesthetist should allow the lung to deflate slowly and only as much as is essential for the surgeon. Any untoward sign, such as gasping respiration, increased cyanosis or cardiac irregularity, should be regarded as an indication to re-expand the lung and attempt to return the patient to optimal condition.

The patient will be subject to an increased degree of anoxia as long as the chest remains open, but the period of maximal danger will be during the establishment of the anastomosis, when the right or left pulmonary artery will be temporarily occluded. Here the speed of the surgeon will be an important factor.

Bradycardia developing during this phase of the operation should be regarded as a sign of extreme anoxia. The slowing of the heart in anoxia appears to be a vagal mechanism which in this instance has a markedly deleterious effect. In the event of anoxic bradycardia, full inflation of the lungs should be carried out at once, and atropine, in approximately one-third the preoperative dose, should be given intravenously to interrupt the vagal mechanism and permit the heart to regain its former effective rate.

At completion of the anastomosis, the patient's color may immediately improve. Oxyhemographic tracings may demonstrate a rise in oxygen saturation from 45 per cent before establishment of the shunt to 80 per cent immediately afterward (20). The condition of the patient should improve during the remainder of the operation. Several other factors, however, remain to complicate the picture.

To recapitulate briefly, the purpose of the operation for tetralogy of Fallot is to increase the flow of blood to the lungs. This is performed by anastomosis of either the subclavian artery, as in the Blalock operation (21), or the aorta, as in the Potts operation (22, 23), to the homolateral branch of the pulmonary artery. Thus, by creation of what amounts to an artificial patent ductus arteriosus, some compensation is made for the narrowed orifice of the pulmonary artery. This does not produce a normal patient, however, for it is to be remembered that the overriding aorta still exists, allowing venous blood to enter the aorta. Furthermore, by creation of the new shunt, an extra burden is added to the work of the heart, for such a shunt may so overtax the heart that cardiac failure will result.

The postoperative period may be extremely perilous. The danger of continued anoxia still exists, and most patients will need oxygen therapy for two or three days. Sedation should be adequate to prevent excessive exertion, but one should not overlook the type of restlessness that is due to anoxia. There will also be danger of thrombosis at the site of anastomosis, and to prevent this it will be necessary to

maintain the blood pressure at a sufficient level to drive a steady flow of blood through the shunt. Necessity for adequate blood volume and blood pressure, and the simultaneous danger of overloading the heart or causing excessive hemoconcentration call for an extreme nicety in gauging the administration of fluids and vasopressor agents. The danger of cardiac failure continues to be a real one for some time during the postoperative period. This failure may come as a result of too large an anastomosis, or from excessive fluid intake. Fluids should be limited to about 30 cc. per pound per day in the early postoperative period (24).

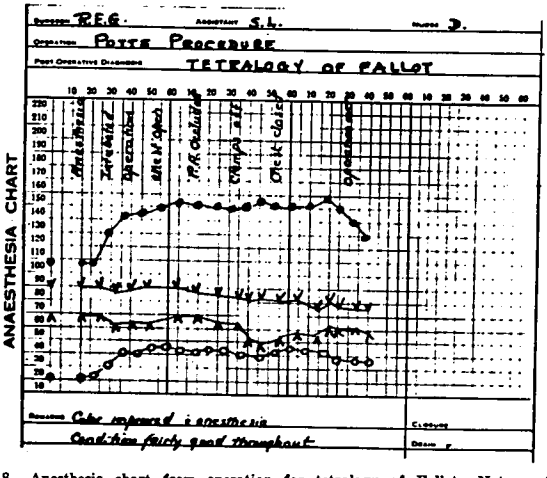


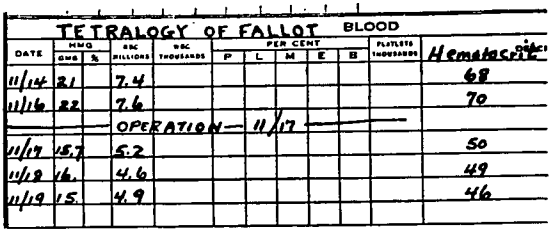

FIG. 8. Anesthesia chart from operation for tetralogy of Fallot. Note rapid pulse, low blood pressure and narrow pulse pressure. In this case there was evidence of establishment of shunt when removal of clamps was followed by widening of pulse pressure.

Mention should be made of those patients who do poorly during operation. Extreme cyanosis, jerky respiration and cardiac slowing may be taken as signs of impending disaster. If oxygenation of these patients does not improve their condition, the anesthetist may feel justified in suggesting abandonment of the procedure. Before doing this, however, he should bear in mind that cyanotic patients bear the insult of thoracotomy poorly unless the surgeon is able to better the patient's condition in some way. While the mortality should be in the range of 8 to 10 per cent in successfully completed cases, that in inoperable or incompleted cases has been almost 50 per cent. Consequently, once the chest is open, the anesthetist should be ready to as-

sume unusual risks if the surgeon sees the possibility of effecting an anastomosis.

The anesthesia chart shows little of especial interest in operations for tetralogy of Fallot (fig. 8). Rapid pulse and respiration are not unusual. The blood pressure and pulse pressure usually remain weak throughout. After anastomosis, there may occasionally appear a widening of pulse pressure, owing to decreased peripheral resistance. The operation requires one and a half to two hours in experienced hands.

The laboratory sheet bears important information and should be watched carefully (fig. 9). Preoperatively, the degree of polycythemia and hemoglobin content indicate the extent of compensation necessary, while the hematocrit reading can be used as a gauge of the likelihood of thrombosis. Hematocrit levels of 80 or more are considered in the critical range.



TETRALOGY OF FALLOT — BLOOD

| DATE | HMG GMS | HMG % | RBC MILLIONS | RBC THOUSANDS | PER CENT P | L | M | E | B | PLATELETS THOUSANDS | Hematocrit |
|---|---|---|---|---|---|---|---|---|---|---|---|
| 11/14 | 21 | | 7.4 | | | | | | | | 68 |
| 11/16 | 22 | | 7.6 | | | | | | | | 70 |
| | | | OPERATION — 11/17 | | | | | | | | |
| 11/17 | 15.7 | | 5.2 | | | | | | | | 50 |
| 11/18 | 16. | | 4.6 | | | | | | | | 49 |
| 11/19 | 15. | | 4.9 | | | | | | | | 46 |

FIG. 9. Laboratory chart of patient operated upon for tetralogy of Fallot. Abnormally high hemoglobin, red cell count and hematocrit decreased abruptly at operation, and remained within normal limits.

Postoperatively, these figures show remarkably rapid alteration. In the accompanying case (fig. 9), erythrocytes, hematocrit, and hemoglobin decreased to normal within three hours after operation and remained there throughout the postoperative course. Failure of the appearance of alteration toward normal may indicate an unsuccessful operative result.

### COARCTATION OF THE AORTA

In patients with coarctation of the thoracic aorta the primary lesion is not difficult to visualize (fig. 10). It is simply a stenosis of the aorta causing what amounts to a complete or high degree of obstruction of that vessel (25). Above the level (cephalad) of the aortic obstruction this block produces hypertension and the development of an extensive collateral circulation (26, 27, 28), while below the level (caudad) of coarctation there results hypotension (29), decrease or loss of arterial pulsation and prolonged circulation time (30).

As will be seen, most of the anesthetic and surgical problems involved in the treatment of these patients revolve about these abnormal factors.

In general, patients with coarctation of the aorta constitute a more hopeful group to deal with than the cyanotic group previously described. The operation is elective, performed prophylactically with the expectation of complete cure of the patient. The operation for tetralogy of Fallot is more in the nature of a palliative procedure. Since the ill effects of coarctation usually do not become manifest until puberty or later, it is not necessary to operate for this disease during infancy or early childhood. This is a decided advantage for the anesthetist. The usual age of these patients at operation is from 10 to 25 years.

As a result of the underlying lesion, patients with coarctation of the aorta have characteristic abnormalities (31). The hypertension in the vessels of the head, upper extremities and upper body may be marked and may give rise to dangerous secondary lesions. Cerebral vascular damage is not uncommon, and arteriosclerosis generally becomes evident at an extremely early age. The increased peripheral resistance causes left ventricular hypertrophy, which in turn may lead to the development of various abnormalities in cardiac conduction mechanism, manifested by electrocardiographic changes such as a prolonged P–R interval or bundle branch block (32, 33). These defects can ultimately result in cardiac failure.

The impaired circulation below the coarctation is evidenced clinically in many patients by retarded development of the lower part of the body and limbs as compared to the upper body and limbs, and in failure of the circulation to respond to increased bodily demands, for example, the sensation of weakness and paresthesias in the legs when the patient climbs a flight of stairs.

The operation for coarctation of the aorta necessitates an extensive incision for exposure (34, 35, 36). After opening the chest, the surgeon must free the aorta at the site of coarctation, place clamps across it proximal and distal to the obstruction, excise the stenosed segment and perform the anastomosis of the free ends of the aorta. If the length of the excised segment is excessive, Gross has been able to bridge the defect with aortic grafts (37). If the origin of the subclavian artery is too near the site of the coarctation, it may be necessary to clamp off this vessel while the aortic anastomosis is being accomplished.

In patients with coarctation, the collateral circulation carries the major part of the blood supply to the lower portion of the body and legs by way of the internal mammary and intercostal vessels. Consequently, vessels of the chest wall are tremendously engorged. The opening of the chest, therefore, may entail considerable loss of blood. It has been found that 500 to 1000 cc. of blood may be lost in adults during the opening of the chest.

The extensive chest wound imposes a handicap on the patient not only through blood loss but also through destruction of an appreciable number of vessels essential in carrying the collateral circulation. Furthermore, after opening the chest, the surgeon may have to sacrifice one or more intercostal arteries at their site of origin in mobilization of the aorta. The effect of this diminished collateral circulation will be an increase in peripheral resistance and, of course, a decreased amount of blood reaching the lower body and legs.

In our experience with these patients there have been two critical points during the operation: namely, the period during which the aorta is clamped off and that immediately following the release of the aortic

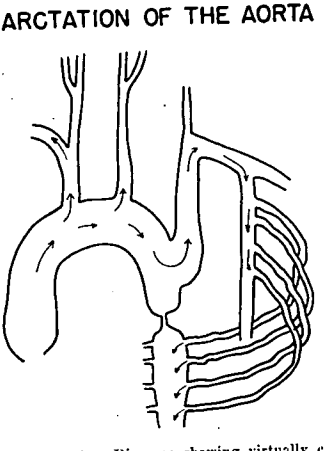



FIG. 10. Coarctation of the aorta. Diagram showing virtually complete obstruction of aorta, with resultant development of huge collateral flow through subclavian, internal mammary and intercostal vessels. It should be noted that direction of flow through intercostal arteries is reversed, that is, from arteries into aorta.

clamps. The placing of the aortic clamps may have widely varying effects on the patients, depending on the position of the aortic clamp. In cases in which the coarctation already produces virtually complete obstruction of the aorta (fig. 10), the placing of a clamp immediately above the obstruction (fig. 11) should cause little or no change in the dynamics. In these cases there may be no change in blood pressure or the blood pressure may rise only 20 or 30 mm. (fig. 12). The total rise takes place within about ten minutes, after which time the blood pressure holds a plateau during excision of the coarctation and resuture of the aorta. This phase may involve half to three-quarters of an hour.

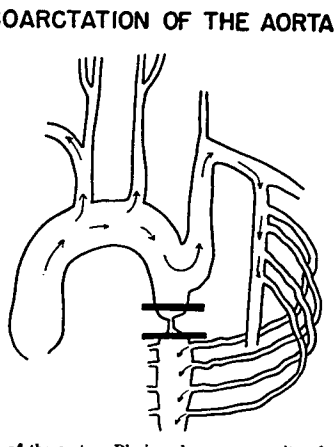



FIG. 11. Coarctation of the aorta. Placing clamps across site of already obstructed aorta produces little change in previously existing conditions.

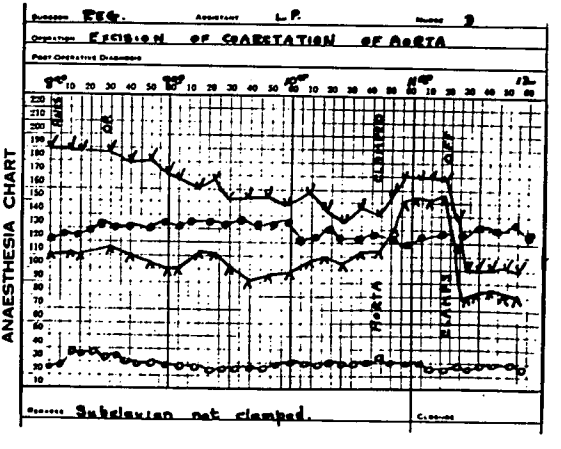



FIG. 12. Anesthesia chart from operation for coarctation of the aorta showing only slight rise in blood pressure produced by placing clamps across site of obstruction.

In addition to the clamping of the aorta, in 15 per cent of the patients operated on by Gross (38) it has been necessary to place a clamp across the left subclavian artery in order to have an adequate proximal segment of aorta for anastomosis (fig. 13). This situation places the patient in a relatively dangerous position, for this added obstruction causes a marked increase in the amount of blood going to the head and a relatively large increase in blood pressure.

If a preoperative level of blood pressure of 180 to 200 mm. is maintained throughout the early part of operation until the time of clamping of the aorta, and if it then happens that the subclavian

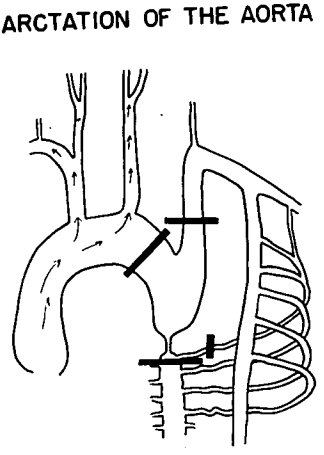



FIG. 13. Coarctation of the aorta. The proximal segment of the aorta is so dilated that the subclavian artery must be clamped in addition to the aorta while correction is performed. This causes great increase in flow to head and right side.

artery must be clamped in addition to the aorta, the arterial pressure may easily rise to as much as 230 or 240 mm. (fig. 14). Such a turn of events has proved disastrous on several occasions. In one patient convulsions developed under these circumstances, probably as a result of excessive cerebrovascular tension. Two other patients had cardiac slowing and arrest, one with complete recovery, the other with temporary recovery and subsequent death. The cardiac slowing can be explained on the basis of the excessive pressure acting through the carotid body and aortic depressor mechanism. Electrocardiograms show evidence of increased vagal activity during this period (39).

Such disturbances have led us to approach the phase of aortic clamping with due concern. In order to prevent the elevation of

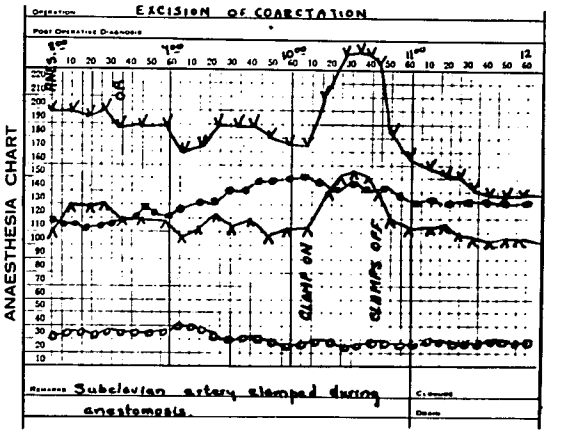



FIG. 14. High elevation of blood pressure following clamping of both aorta and subclavian artery. The blood pressure was 175 mm. before clamping of vessels, and the resultant 65 mm. rise of 65 mm. brought pressure to a dangerous level of 240 mm.

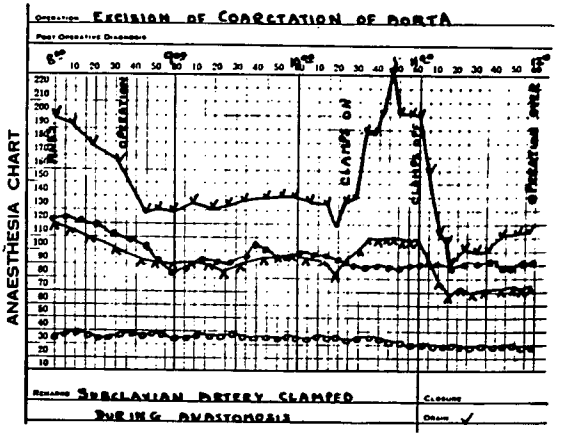



FIG. 15. Anesthesia chart showing 90 mm. rise in systolic pressure following clamping of both aorta and subclavian artery. The danger of reaching critical blood pressure levels was prevented by allowing the pressure to fall to 140 mm. before the vessels were occluded.

blood pressure to excessive heights, our principal aim has been to establish the blood pressure level at about 150 mm. prior to occlusion of the aorta. This can be done by balancing the blood lost against that replaced, and usually means that the original blood pressure level must be allowed to fall gradually until the desired level is reached (fig. 15). The replaced blood will consequently be less than that lost up to that point. With the blood pressure at this reduced level, a pressure rise of 70 to 80 mm. will still not raise the total pressure to extreme limits. The rise in blood pressure occasioned by placing of clamps will reach a maximum within ten to fifteen minutes. Any deficit in blood replacement should then be made up gradually, in preparation for release of clamps and tremendous reduction of peripheral resistance (fig. 16).

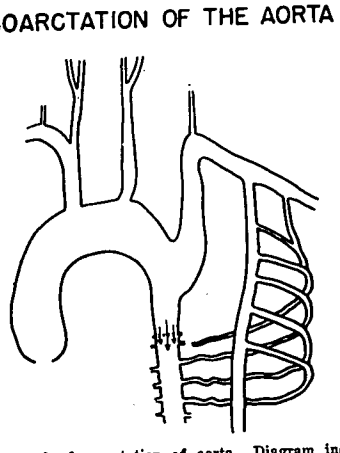



FIG. 16. Effect of removal of coarctation of aorta. Diagram indicating re-establishment of normal flow through aorta, with resultant decrease in peripheral resistance.

In all patients, release of the aortic clamps entails the danger of profound shock. The first patient in this series died suddenly upon release of the aortic clamps (fig. 17), and similar fatalities have occurred at other institutions. These have, undoubtedly, been the result of either too rapid release of the clamps, an inadequate circulating blood volume at that time or a combination of both factors. Two measures of cardinal importance should be observed: namely, *very gradual release of the aortic clamps and complete replacement of blood volume.* With slow release of clamps in the presence of an adequate blood volume, the blood pressure will fall to a level slightly below that present before the clamps were placed. It may then fall gradually to about

110 mm. which is probably a desirable level. Strict adherence to these simple principles will enable the patient to withstand procedures of considerable magnitude without mishap (fig. 18).

Proper blood replacement is unquestionably the largest problem for the anesthetist during these procedures. The total amount of blood required in full grown patients may be surprisingly large. It is not unusual to use 3 liters in uneventful cases, while as much as 13 liters have been required in the presence of copious hemorrhage. In the event of sudden hemorrhage which requires rapid blood replacement, inaccurate methods of measuring blood loss may leave one in doubt as to whether too much or too little blood has been administered. A

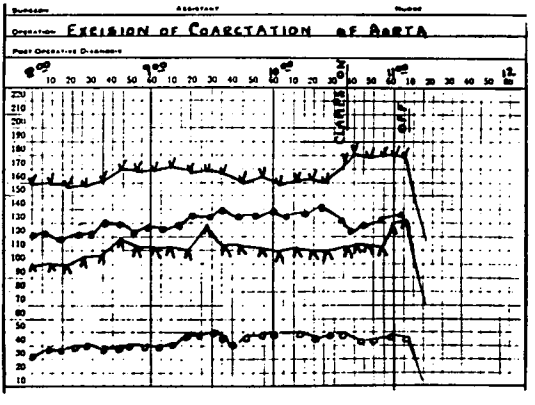



FIG. 17. Effect of sudden decreases of peripheral resistance at time of removing clamps in operation for excision of coarctation of aorta. Inadequate blood replacement and sudden removal of clamps caused death of the patient.

practical method of determining blood volume would be of tremendous value in thoracic surgery. The present methods utilizing Evans blue dye (40), and so forth, are too time-consuming to be of use during operation, but may give delayed information. The measurement of venous pressure, as suggested by Lundy (41), probably deserves more clinical application in such cases.

In our cases blood loss is measured by using dry sponges of known weight and weighing them after use to determine the blood absorbed. A scale has been calibrated for this use (42). Blood in the suction bottle is also measured throughout the operation. Although blood loss on gowns and drapes cannot be measured accurately by this method, the measurement of sponged and suctioned blood at repeated

intervals has been of great assistance in the estimation of replacement needs.

In a total of 155 operations for coarctation of the aorta performed by Gross there has been a mortality of 8.5 per cent. In reviewing the fatal cases it has been shown that the deaths, almost without exception, were the result of excessive hemodynamic strain superimposed upon an already impaired heart.

Cardiac arrhythmias were observed with surprising infrequency in this series of operations upon the great vessels. The fact that the pericardium and heart were not molested played a large part in the avoidance of conduction disturbances, while the use of ether as the principal anesthetic agent also may have contributed.

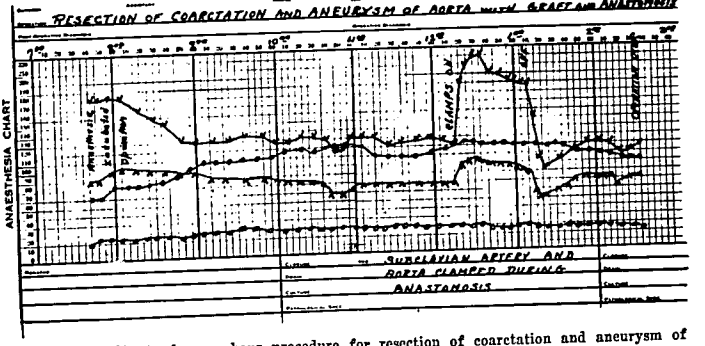



FIG. 18. Chart of seven hour procedure for resection of coarctation and aneurysm of aorta with graft and anastomosis. In this operation it was necessary to clamp both aorta and subclavian artery during anastomosis. Careful control of the blood pressure level allowed a rise of 100 mm. in the systolic pressure after placing of clamps. Replacement of blood volume and slow removal of clamps prevented dangerous drop in blood pressure on removal of clamps. Total of 5500 cc. of blood was administered. Condition of patient was excellent throughout the procedure.

No cases of ventricular fibrillation or of apparent increased myocardial irritability were encountered. The few conduction disturbances that were seen appeared only in the presence of such markedly unphysiologic conditions as acute heart strain, severe anoxia, or excessive changes in blood pressure. Treatment of these disturbances was centered in the correction of the underlying abnormal conditions rather than in the use of pharmaceutical agents.

During operations on cyanotic patients bradycardia was the principal type of cadiac irregularity. As has been mentioned, and as reported by Ziegler (43), this appears to be of vagal origin in the presence of anoxia. It is best treated by oxygenation and administration of atropine. Our experience has also led us to agree with Ziegler

that in the presence of an arrested nonfibrillating heart, epinephrine is the most effective agent to initiate the cardiac impulse.

Serial electrocardiographic tracings taken during operations for coarctation of the aorta have revealed only minor abnormalities as long as physiologic conditions existed. Moreover, no marked alterations in conduction occurred following the placing or releasing of aortic clamps with moderate changes in resistance. Excessive hypertension brought evidence of increased vagal action and cardiac arrest. Arrest occurring upon rapid release of aortic clamps undoubtedly is due to sudden disappearance of effective blood volume.

In general, it has been thought that use of drugs for prevention of cardiac arrhythmias should be limited to cases showing definite conduction disturbances before operation. In such instances quinidine or pronestyl has been used, depending on individual indications.

## SUMMARY

An attempt has been made to outline the chief anatomic and physiologic abnormalities in patients with patent ductus arteriosus, tetralogy of Fallot and coarctation of the aorta, and to explain the special problems encountered in each type of patient.

Patients with patent ductus arteriosus have a recirculation of oxygenated blood through the pulmonary circulation, resulting in left ventricular strain and danger of cardiac failure. They are completely cured by operation.

Patients with tetralogy of Fallot have a recirculation of unoxygenation blood through the systemic circulation, resulting in right ventricular hypertrophy, cyanosis and polycythemia. Operation offers only partial relief from anoxia and thrombosis, and adds further danger of postoperative cardiac failure.

Patients with coarctation of the aorta are cured by operation, but only at the cost of a formidable procedure involving extensive blood loss and hemodynamic changes.

Hazards to be encountered during operations for congenital cardiac disease are principally the result of altered hemodynamics and anoxia, while cardiac irritability and arrhythmias play a relatively insignificant role.

All operations upon the great vessels necessarily involve the risk of sudden, profuse hemorrhage.

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